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NOTES OF A CASE OF
DERMATITIS HERPETIFORMIS
DURING THIRTEEN YEARS.

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THE notes here presented, portraying the disease I have named dermatitis herpetiformis, are of such interest that they are given in full, with the view of directing attention to the many and peculiar phases this remarkable disease may assume. The long period the disease was under observation and the extended notes, which were made at the time, give us valuable information on the natural course of the disease—a point always of importance in the study of cutaneous affections:

H. H. McK., aged thirty-one, an American, and a painter by occupation, was admitted to the Philadelphia Dispensary for Skin Diseases, Philadelphia, July 6, 1874. He was in impaired health, and had been suffering from the skin disease for which he sought relief since 1871—a period of three years. He was of small frame and spare, and was apparently in a state of nervous exhaustion, being weak, tremulous, dyspeptic, and despondent about his condition. He was married, and was temperate in his habits.

The following clear history was obtained : In 1871 the first disease of the skin ever experienced manifested itself in the form of lesions which he describes as "hives." They came unexpectedly and suddenly, and occupied every portion of the surface except the face, being particularly numerous and thickly set about the elbows and knees. They were erythematous in character, and were split-pea and finger-nail sized, raised, and flat. Some were discrete, but the greater number were confluent. The whole surface was pretty well covered with the efflorescence, and was the seat of violent itching. The attack is described as having possessed all the features of urticaria. It gradually subsided in the course of three or four days, when a crop of small, rounded, flat *pustules* began to appear here and there, especially about the knees and elbows, accompanied by itching. They increased slowly in size, and crusted with a brownish, adherent crust. A number of them came out, varying in size from a split pea to a small finger-nail. They all ran a similar course, lasting from five or six days to a fortnight. By the end of four weeks they had completely disappeared, but, almost before the crusts of the earlier lesions had fallen off, a crop of *vesicles* and *blebs*, of all sizes and shapes, suddenly appeared, distributed quite generally over the extremities and upon the face. There were great numbers of them upon the face, and they varied in size here from a pin-head to a hazel-nut. Upon the general surface they were mostly small—pin-head and split-pea sized. Some were raised and semi-globular in form; others were flat. The lesions, as stated, were *vesicles* and *blebs* having perfectly clear contents, and were without areolæ. They were very itchy. There were no signs of *pustules*.

This attack lasted about two months, new vesicles and blebs appearing from day to day, and gradually subsided. The patient speaks of the experience as having been very trying, not only on account of the presence of the lesions and the discharge and crusting, but also because of the itching.

For several months he remained comparatively well, when a copious mixed eruption—composed of small *vesicles*, *pustules*, *vesico-pustules*, and *vesico-papules*, accompanied with itching—made its appearance upon the regions previously affected. This

was the first occasion of the lesions being distinctly multiform. This attack lasted only a fortnight, vanishing quite suddenly, and leaving the skin in a better state than for months. This last form of eruption, however, soon recurred, lasting, as previously, but a few weeks, and, as before, leaving him apparently quite well. He has had many attacks of this character, at intervals of a few months, up to the present time. The disease has thus continued its course, uninfluenced by the varied treatment to which he has been subjected. He does not think that the seasons have at all influenced the natural course of the disease.

It was at this period (July, 1874) that the patient first presented himself to me. He showed the pustular phase of the disease, which had but recently made its appearance. There were about a score of disseminated, split-pea and finger-nail sized, flat, mostly crusted with a yellowish-brown, adherent, flat, ecthyma-form crust, here and there distinctly clustered *pustules*. They occupied the lumbar and sacral regions, the buttocks, and the elbows and knees. They had appeared a fortnight previously, and had been and were still accompanied by itching. The more recent lesions I observed were surrounded with bright-red, highly inflammatory areolæ, and were peculiar in that they inclined to crust in their center and to spread on the periphery, forming more or less distinct, narrow rings of pustulation immediately beyond the line of the crust. This feature was not constant, but existed in connection with a number of the lesions, and constituted a symptom which at once arrested my attention.

The case was regarded as an unusual one, the form of disease being entirely new to me, and this observation was so recorded in connection with the notes. The lesions bore a close resemblance to cachectic ecthyma, but the localities attacked were those seldom invaded in this latter disease. The pustular variety of eczema, aggravated by some irritating application, also suggested itself, as did, moreover, impetigo contagiosa. The contents of several of the pustules, as well as the crusts, were at the same time submitted to microscopical examination with a view of determining if any parasite was present. The

patient was placed upon a saline laxative mixture, together with full doses of the tincture of chloride of iron. No local treatment was ordered. The case remained under observation three months, and, according to the notes recorded, the patient was in about the same condition at the expiration of this period as upon admission; the skin, however, underwent several decided changes during the three months, having been on one occasion almost well, but the lesions soon reappeared. The disease did not seem to be in the least degree influenced by treatment, whether internal or local, new pustules, small and large, appearing from week to week. In November, 1874, he ceased attending the dispensary, much discouraged. The diagnosis was as obscure at this time as on admission.

In January, 1879, four years after the foregoing note, he presented himself for treatment at the Hospital of the University of Pennsylvania. I at once recognized my former patient of the Dispensary for Skin Diseases. He stated that he was still afflicted, but that until recently it had existed in a milder form, and that he had experienced intervals of from one to six months of comparative freedom, but he had never been entirely free of eruption. The general health had failed considerably, the nervous system had become more disturbed, and he was habitually constipated. During the four years he had employed various internal and local remedies, but without benefit. The general course of the disease had been about the same as when under observation four years before. He had noted four kinds of lesions, which would manifest themselves together or in different attacks of eruption; these were true *pustules*, containing from the beginning a whitish puriform fluid, and of variable size, from a pin-head to a silver quarter-dollar; *vesicles*, containing a clear yellowish fluid, likewise of variable size; *blebs*, varying in size from a split pea to a half-cherry; and, lastly, *papulo-vesicles*, acuminate in form, and exuding a small amount of a serous, gluey, gummy product. The "watery blisters," as he termed the vesicles and blebs, had, on every occasion of their appearance, been very itchy; the pustules itched but little. The vesicles and blebs grew rapidly, reaching their full size in a few days, and were accompanied by but slight inflammation about

their bases; the pustules, on the other hand, were always markedly inflammatory. On each occasion of the pustular form of eruption he "felt badly," and was in poor general health. He had had but three or four "pustular attacks," the rest having all been vesicular and bullous. The disease, whatever the form of lesion, always manifested itself in attacks, which would announce themselves at intervals of weeks or months, one scarcely subsiding before another would appear. He does not remember that any of the attacks were preceded by chills or fever. The seasons seemed to exert no influence on the course of the disease. He noticed that frequently the lesions inclined to come out in the form of groups or clusters, four or five appearing within a radius of an inch or two, and often coalescing. The regions invaded have been the same as in the beginning—the face, neck, elbows, knees, sacral region, and buttocks being the favorite localities. The genitalia, hands, and feet have generally been free; the palms and soles have always escaped.

His condition when I saw him in January, 1879, was very similar to that recorded as existing when I first observed the case in 1874, and therefore need not be described in detail; so much did his condition resemble the former state, as regards the general character and distribution of the eruption, that it seemed as though the very same lesions were present. The pustules were the only lesions. They were of all sizes, most of them being as large as dimes and quarter-dollars; rounded or irregular in shape; crusted with yellowish-brown crusts; flat; here and there clustered or about coalescing; in all stages of development, and surrounded by highly inflamed, extended, bright-red areola. They, moreover, possessed the characteristic before noted of extending around the periphery and just beyond the line of the central crusting. The patient was ordered wine of iron, with three-minim doses of liquor potassii arsenitis thrice daily. With the view of observing the course of the lesions, local treatment was withheld.

March 11, 1879.—The case has been under constant observation since the previous note, six weeks ago, during which time the disease has behaved much as it did on the first occasion I had of studying it. The lesions have pursued a rather slow

course, considering their highly inflammatory nature, an average pustule lasting, from the beginning until the crust dropped off, from two to three weeks. Many new lesions, all strictly pustular in character, have from time to time appeared, as a rule, in crops, while the older ones have, at the end of a variable period, disappeared, leaving persistent dark-red, brownish, pigmented marks. No scars remain. One peculiarity of mature pustules has not been referred to, namely, the tendency for the inflamed skin around the crust to pucker, giving the skin a wrinkled, drawn-up, glistening appearance. The disposition of the lesions to group has always been present, though at times only to a slight extent. It has not, however, been observed with all of the lesions, many appearing singly. As they increased in size there would generally be noted a number of discrete or confluent small, pin-head sized, flat pustules immediately around the circumference of the original lesion in the form of a ring. This feature varied, being much more pronounced in some cases than in others, and at times was wanting. The pustules, however, seemed to grow in this manner, a new ring of minute pustular points springing up every few days around the crust until the lesion reached its determinate size. The crusts were adherent; upon lifting them up, a superficial, reddish excoriation, covered with a slight puriform fluid—as in the case of ecthyma—presented itself. Little or no bleeding occurred. Itching has not been severe, but the lesions have “felt sore.”

Three nights ago he was attacked with violent itching of the general surface, accompanied with an efflorescence which he describes as having been identical with certain attacks before experienced on several occasions, and which was plainly urticarial in nature. It began, while in bed, upon the arms, but soon extended itself over the whole surface except the hands and feet, the backs, as well as the palms and soles, remaining free. The eruption was *erythematous* and very red, and exceedingly itchy; so much so that he was kept awake all night. The next day the eruption disappeared, but he felt poorly and complained of loss of appetite and malaise. Last night, just after going to bed, it suddenly reappeared in the same manner as two days before. The attack was preceded by chilliness and malaise;

later he perspired profusely. To-day he feels better, and the rash is leaving the surface. The skin, however, is still red; rough in patches; slightly papular and vesicular, and has the appearance on the arms of an erythema multiforme in the stage of declination; scratch-marks abound. It seems as though the attack had been abortive, the development of the papular and vesicular lesions having been checked early in their course.

16th.—During the past five days the eruption has been disappearing, with slight desquamation and considerable pigmentation, some of the patches on the trunk somewhat resembling irritated tinea versicolor. Within the week a half-dozen, large and small, *pustules* have come on the arms and buttocks, identical with those which have been described.

25th.—A number of variously sized pustules have been appearing here and there over the surface, in particular on the neck, around the navel, on the lumbar and sacral regions, and over the buttocks and thighs. They all incline to dry and crust in the center, sometimes as a depressed, cup-shaped crust, and to spread on the circumference, thus extending their size. Their edges are more or less “puckered” and slightly raised; their areolæ large, dark-red in color, and contracted, presenting a somewhat stellate, radiate appearance. There is now but little itching. He is still taking four minims of the liquor potassii arsenitis, which he has been using for a month, but with no benefit.

June 29th.—Has been annoyed greatly with disease since the last note, which has manifested itself, as heretofore, in the form of distinct outbreaks appearing at irregular intervals. But at no time within the past three months has he been entirely free. The lesions at present are *vesicular* and *bullous*, from the size of large pin-heads to coffee-grains, and are scattered especially over face, arms, and forearms, and over the region of the spinal column. They are accompanied with intolerable itching. During the last six weeks he has been using half-ounce doses of linseed-meal, with the view of improving the nutrition, but without benefit.

June 1, 1884.—Five years have elapsed since the last note, during which period I have not seen the patient until a few

days ago. He is still suffering with the disease, having never obtained any relief from the manifold treatment to which he has been subjected. For the past three or four years the attacks have been lighter than formerly, and the lesions for the most part vesicular in character. At the present time the face, neck, and trunk are the seat of small, ill-defined vesicles and papulo-vesicles, accompanied, as before, with itching. The general health is improved, and he is able, when not suffering from an attack, to attend to his daily duties.

I shall make no comments upon the case except to say that it illustrates a typical expression of this peculiar disease, characterized as it is by rare multiformity. It shows almost all of the lesions to which I have called attention in describing the disease,* and upon which its several varieties have been based. In conclusion, I may add that the affection is usually well defined, having distinctive features which enable it to be readily recognized, although it may be confounded with eczema, herpes, and pemphigus, according to the lesions at hand.